



(CASE REPORT)



## Adenocortical carcinoma: A rare malignant tumor

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### Abstract

Adrenocortical adenocarcinoma (ACA) is a rare malignancy that develops at the expense of the adrenal cortex. The key to its treatment lies in surgery, with complete resection of a localized form offering the best chance of cure. This article presents the case of a 68-year-old man treated for adrenocortical adenocarcinoma at the Hassan 2 University Hospital in Fez. The patient, with no notable pathological history, consulted for dizziness and abdominal pain, leading to the discovery of a malignant adrenal mass on radiological evaluation. A successful left laparotomy adrenalectomy was performed, confirming, by pathological analysis, a Weiss score 4 adenocarcinoma. Immunohistochemical results showed expression of cytokeratin AE1/AE3, while other markers were absent. The article highlights the efficacy of surgery in the treatment of localized forms of this rare tumor, while highlighting diagnostic challenges and the need for international coordination in adrenocortical adenocarcinoma research and care.

**Keywords:** Adenocortical Carcinoma; Case Report; Malignant Tumor; Adrenal Cortex; Surgery; Laparotomy

### 1. Introduction

Adrenocortical carcinoma is a primary rare malignant tumor developed at the expense of the adrenal cortex. Surgery is the key, since "complete" removal of a localized form is the best chance of a true cure. Anatomopathological analysis remains the cornerstone of the diagnosis of adrenal tumors, and of CS in particular. We will discuss a rare case of a 68 years old adrenocortical carcinoma treated in the CHU Hassan 2 in Fes .

### 2. Clinical case

The patient was 68 years old.

With no notable pathological history

The history of the disease dates back to 2 months prior to admission, when the patient began to experience dizziness and abdominal pain, prompting a consultation with a private physician, who found an abdominal mass and referred him to us for further treatment.

On clinical examination

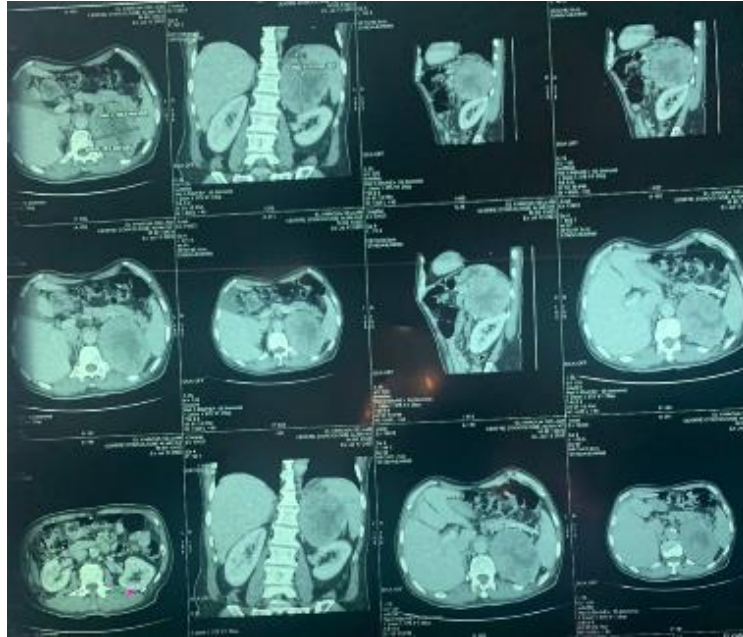
The patient was conscious, normotensive, apyretic

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With a soft abdomen, presence of left lumbar tenderness with left lumbar contact

With No signs of feminization or Cushing syndrome.

Biological test showed following results: Hb : 14,8 , GB : 5K , Creat a 7 , Na+ 137 , K+ 4,3 Glycemia 1,06; Measurement of metanephrens was negative; The cortison and also the dexamethasone suppression test was negative.



**Figure 1** CT SCAN images revealing a large left adrenal mass with heterogeneously enhanced tissue density delimiting multiple necrotic zones measuring 100\*78\*91 mm

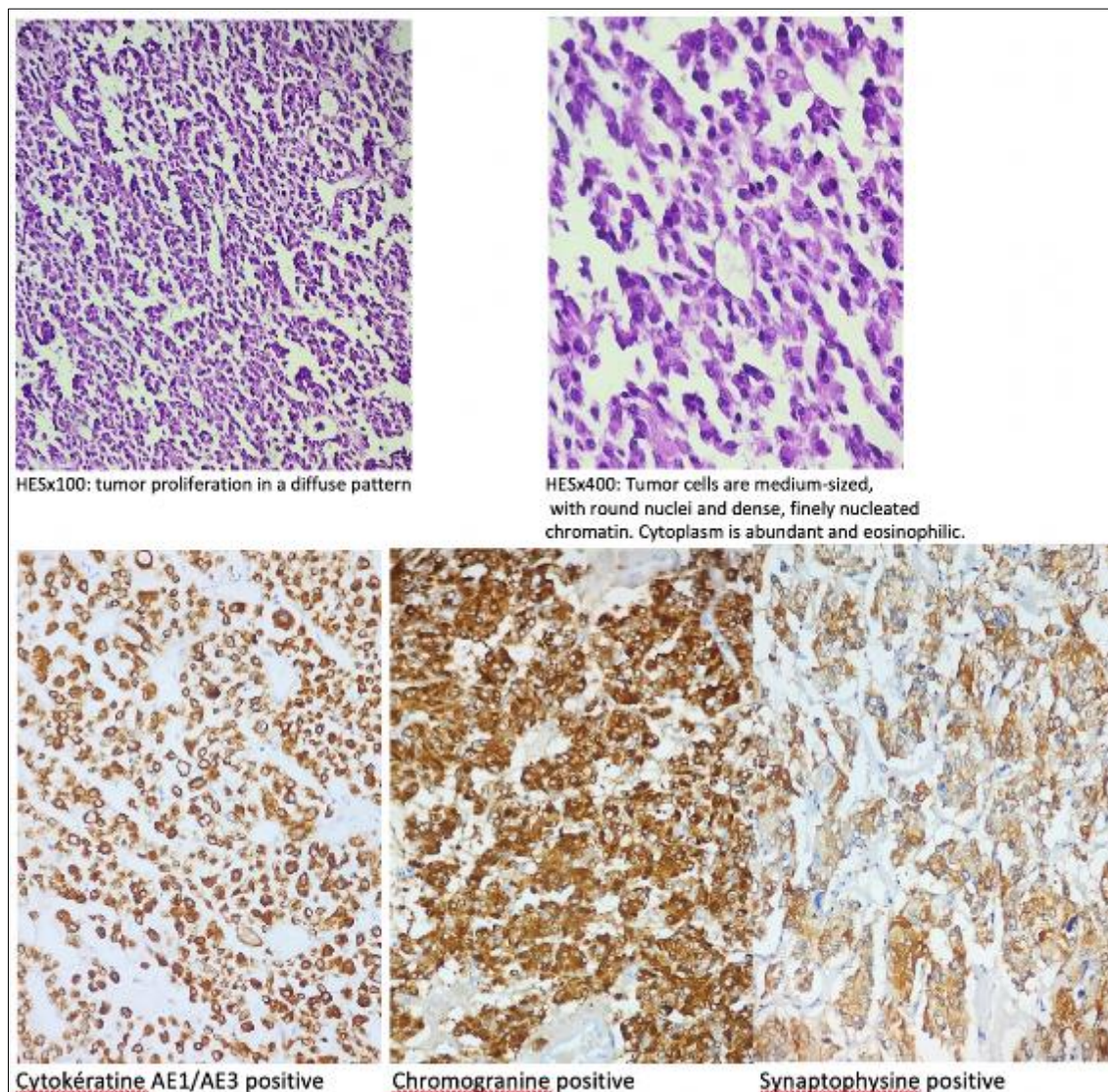


**Figure 2** The adrenalectomy piece after surgery

A thoracic-abdominal-pelvic CT scan revealed a large left adrenal mass with heterogeneously enhanced tissue density delimiting multiple necrotic zones measuring 100\*78\*91 mm with absolute wash out at 10%, the mass comes into intimate contact with the upper pole of the left kidney without a separating fat border classified T4N0M0 according to TNM, and stage 3 according to the ENS@T classification. (Figure1)

The patient underwent a left laparotomy adrenalectomy, during which the mass was not adherent to the kidney or adjacent organs, and the postoperative course was straightforward. (Figure 2)

The anatomopathological and immunohistochemical study came back in favor of a Weiss score 4 adrenocortical carcinoma, in fact the tumor cells expressed cytokeratin AE1/AE3, they did not express cytokeratin 7, 20 l carbonic anhydrase 9, vimentin, CD 10 PS100 CD117 l inhibin chromogranin, synaptophysin and TTF1.(Figure 3)



**Figure 3** Pictures of the anatomopathological and immunohistochemical study.

### 3. Discussion

Adrenocortical carcinoma is a primary malignant tumor developed at the expense of the adrenal cortex, defined by a Weiss score  $\geq 3$ .

It is a rare tumor (1 or 2 cases per million per year), with two peaks of occurrence, in the first decade and between the ages of 40 and 50. (1)

The main circumstances of discovery are:

- -Signs of steroid hormone hypersecretion: Cushing's syndrome, hyperandrogenism in women, hyperestrogenism in men, hypertension with hypokalemia.
  - Tumor syndrome (palpable mass, distant metastasis)

Our patient presented with only an abdominal pain and a palpable mass on the left.

Biologically, steroid and precursor hypersecretion is systematically investigated with metanephrine and normetanephrine assays and a cortisol minute suppression test (1). A general check-up will assess also: Glycemia, blood count, blood ionogram (kalemia), kidney and liver functions.

In our case, the biological check-up came back without any abnormalities, no secrecy.

2 Radiological examinations are essential: CAT scan and FDG PET scan. Abdominal MRI is a 2nd-line examination. They enable staging c TNM and assessment of respectability, as well as arguments in favor of malignancy.

On CT Most CS have a diameter greater than 5 cm at diagnosis, are heterogeneous with irregular margins, spontaneous density > 10 HU with wash-out < 50% after injection of contrast (2)

Positron emission tomography (PET) with 18-fluoro-deoxyglucose (18 FGD) helps to distinguish benign lesions from malignant tumors, and assists in the search for distant metastases, which are sometimes silent and only revealed by this examination. FDG-PET is also useful for monitoring treated patients (3).

The CT data in our patient was in favour of the malignant origin of the mass, the PET scan was not performed due to the unavailability of the product at the hospital.

Surgery is the key, since "complete" removal of a localized form is the best chance of a true cure. Over the past five years, there has been debate about the best surgical approach (4). Laparotomy remains the standard approach. Laparoscopic adrenalectomy is considered only in very select cases.

Conventional radiotherapy, administered as adjuvant therapy can extend the recurrence-free time (5) . However, a recent American study does not seem to confirm these data (6).

The real benefit of adjuvant treatment with mitotane in all patients with CS remains to be discussed, particularly for those with a "low" or "intermediate" risk of relapse (i.e. Ki-67 10%, R0 and absence of metastasis)

In our case the patient benefit from a laparotomy removal of the mass, no adjuvant therapy was proposed due the the low risk of relapse and the luck of available adjuvant therapy data

Anatomopathological analysis remains the cornerstone of the diagnosis of adrenal tumors, and of CS in particular. However, it remains a real challenge, even for the most experienced anatomopathologists, for two main reasons. The first difficulty is to establish with certainty the adrenocortical origin of the lesion. For this, steroidogenic factor 1 (SF-1) immunostaining appears to be the most sensitive and specific marker (7). The second difficulty is discriminating between benign and malignant lesions. The most widely used score is the Weiss score, which comprises 9 different items (8):. It is accepted that a score of 3 or more is more than likely indicative of the malignant nature of the tumour.

Numerous parameters have been studied. One criterion seems to be emerging: tumor grade, assessed by counting mitoses in the tumor and/or by Ki-67 immunohistochemistry, which quantifies the proportion of cells in mitosis. High-grade tumours appear to be associated with a poor prognosis (8).

Several molecular markers also appear to be associated with a poor prognosis, and are measured by immunohistochemistry (nuclear accumulation of p53, high intensity of SF-1 labelling, nuclear accumulation of beta-catenin).

Our anatomopathological and immunohistochemical study came back in favor of a Weiss score 4 adrenocortical carcinoma, the mitotic count found 8 mitoses/50 without necrosis or emboli, he did not show the signs of a poor prognosis in IHC.

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#### 4. Conclusion

In conclusion, this case of adrenocortical carcinoma highlights the effectiveness of laparotomy adrenalectomy in treating localized forms of this rare tumor, supported by a detailed pathological analysis. Immunohistochemical results aided tumor characterization despite diagnostic challenges, emphasizing the need for advanced techniques.

Since it is a rare disease, it is absolutely essential that research and patient care be organized on a national and international level.

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### **Compliance with ethical standards**

#### *Disclosure of conflict of interest*

No conflict of interest to be disclosed.

#### *Statement of informed consent*

Informed consent was obtained from all individual participants included in the study.

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### **References**

- [1] Tabarin A, Bardet S, Bertherat J, Dupas B, Chabre O, Hamoir E, Laurent F, Tenenbaum F, Cazalda M, Lefebvre H, Valli N, Rohmer V. *French Society of Endocrinology Consensus. Ann Endocrinol (Paris). 2008 Dec;69(6):487-500.*
- [2] WFJr., Young. Conventional imaging in adrenocortical carcinoma: update and perspectives. *Horm Cancer* 2011;2:341-7.
- [3] Deandreis D, Lebouilleux S, Caramella C, Schlumberger M, Baudin E. FDG PET in the management of patients with adrenal masses and adrenocortical carcinoma. *Horm Cancer* 2011;2:354-62.
- [4] Gaujoux S, Bertherat J, Dousset B, Groussin L. Laparoscopic adrenalectomy for adrenocortical carcinoma: a medico-surgical perspective. *Ann Endocrinol (Paris)* 2012;73:441-7.
- [5] Fassnacht M, Hahner S, Polat B, Koschker AC, Kenn W, Flentje M et al. Efficacy of adjuvant radiotherapy of the tumor bed on local recurrence of adrenocortical carcinoma. *J Clin Endocrinol Metab* 2006;91:4501-4.
- [6] Habra MA, Ejaz S, Feng L, Das P, Deniz F, Grubbs EG et al. A retrospective cohort analysis of the efficacy of adjuvant radiotherapy after primary surgical resection in patients with adrenocortical carcinoma. *J Clin Endocrinol Metab* 2013;98:192-7.
- [7] Sbiera S, Schmall S, Assie G, Voelker HU, Kraus L, Beyer M et al. High diagnostic and prognostic value of steroidogenic factor-1 expression in adrenal tumors. *J Clin Endocrinol Metab* 2010;95:E161-71.
- [8] Lau SK, Weiss LM. The Weiss system for evaluating adrenocortical neoplasms: 25 years later. *Hum Pathol* 2009;40:757-68.